

Cross sectional echocardiographic feature in carcinoid heart disease

A mechanism for tricuspid regurgitation in this syndrome

M K DAVIES, P J LOWRY, W A LITTLER

From the Department of Cardiovascular Medicine, East Birmingham Hospital, Birmingham

SUMMARY In a patient with severe tricuspid regurgitation and mild pulmonary stenosis due to carcinoid heart disease cross sectional echocardiography showed nodular thickening and coaptation of the tricuspid leaflets at the beginning of systole. The leaflets were, however, seen to be increasingly pulled apart as right ventricular systole proceeded. This finding, which is probably due to traction on the leaflets by the thickened chordae tendineae, is therefore a mechanism of valvular incompetence, perhaps accounting for the particularly severe tricuspid regurgitation seen in carcinoid heart disease.

The carcinoid syndrome is a rare cause of acquired cardiac disease causing predominantly right sided lesions.¹⁻³ Cardiac involvement is a frequent cause of death, the commonest valvular lesions being a combination of tricuspid incompetence and stenosis with pulmonary stenosis with or without pulmonary incompetence. The advent of cross sectional echocardiography as a non-invasive method of diagnosis has led to recent reports delineating the echocardiographic features of the carcinoid syndrome.^{4,5}

We report a case of carcinoid heart disease in which a new echocardiographic feature accounted for the gross tricuspid incompetence often seen in the syndrome.

Case report

A woman aged 48 years was admitted with a six month history of increasing leg oedema, dyspnoea on exertion, general lethargy, and non-specific chest pain. On examination she had a sallow complexion and a malar flush. Her jugular venous pressure was noticeably raised with prominent v waves. She was in sinus rhythm and normotensive. Clinically she had a parasternal heave, and on auscultation murmurs of predominantly tricuspid incompetence with perhaps some stenosis were audible. A pulmonary ejection systolic murmur was also heard. The liver was enlarged and was palpable 4-5 cm below the costal margin but

was not obviously pulsatile. She also had pronounced bilateral oedema of the legs. Routine haematological and biochemical investigations including liver function tests were normal apart from a normochromic normocytic anaemia (haemoglobin 11.7 g/dl). Her electrocardiogram showed generalised reduction in voltages.

M mode echocardiography showed a large right ventricle with paradoxical septal motion consistent with right ventricular volume overload. The cross sectional echocardiogram (Fig. 1) was diagnostic of carcinoid.^{4,5} The right atrium and ventricle were dilated. The tricuspid valve leaflets showed nodular thickening and were coapted at the end of diastole and the beginning of systole. During right ventricular systole, however, while the right ventricular cavity was diminishing, this coaptation between the leaflets was lost and the tricuspid valve opened, reaching a maximum after mid-systole. The septal leaflet was immobile and foreshortened. The mitral and aortic valves were normal, and the pulmonary valve could not be visualised.

Urinary 5-hydroxyindole acetic acid concentrations were noticeably raised at 330-555 $\mu\text{mol}/24\text{ h}$ (normal $<20\text{ }\mu\text{mol}$). Liver ultrasonography showed gross hepatic enlargement with large tumour deposits in both lobes. This finding was confirmed at laparoscopy, the liver biopsy specimen showing a carcinoid tumour.

Cardiac catheterisation and right ventricular angiography confirmed severe tricuspid regurgitation (right atrial "v" wave 22 mm Hg) with mild pulmo-

Requests for reprints to Dr M K Davies, Department of Cardiovascular Medicine, East Birmingham Hospital, Bordesley Green East, Birmingham B9 5ST.

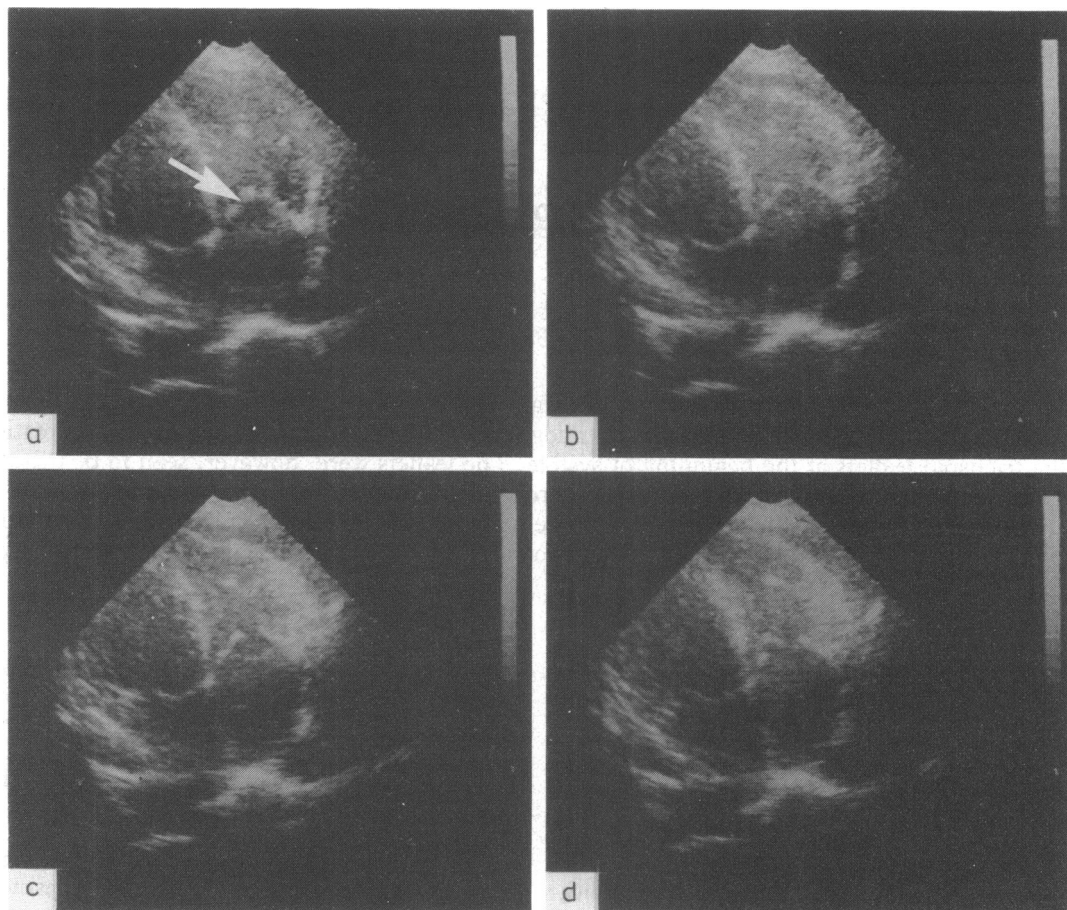


Fig. 1 Cross sectional echocardiogram in four chamber view showing consecutive systolic frames (right ventricle on right of each frame). At the beginning of systole (a) the right ventricle and right atrium are enlarged compared with the left ventricle and left atrium. The tricuspid valve is irregularly thickened and has a high reflectance compared with the mitral valve but the leaflets are coapted. As systole proceeds and the right ventricular cavity dimensions decrease (b, c) the tricuspid valve leaflets lose their coaptation and the tricuspid valve opens to reach a maximum at end systole (d). The septal leaflet is relatively immobile (a, arrow). The mitral valve is normal and remains closed throughout systole.

nary stenosis (peak systolic gradient across the pulmonary valve of 10 mm Hg) and normal left heart pressures. Shortly after these procedures the patient underwent a successful tricuspid valve replacement using an Abrams valve. At operation the tricuspid valve leaflets were found to be thickened and retracted (Fig. 2).

Discussion

Many of the recently described cross sectional echocardiographic features of carcinoid heart disease⁴⁻⁶ were seen in this patient. Firstly, the tricuspid valve leaflets were thickened in a nodular fashion and had a high reflectance. Secondly, the septal leaflet was

immobile and foreshortened, and the mitral valve was normal, suggesting that the right valvular lesions were not due to rheumatic heart disease. Unlike all previous reports, however, in which the tricuspid valve leaflets were seen to be fixed in a semi-open position throughout the cardiac cycle, the tricuspid valve leaflets were quite clearly coapted at the end of diastole in our patient. Indeed, the mechanism of the gross tricuspid regurgitation was clearly seen, with the leaflets gradually opening and the valve orifice increasing throughout right ventricular systole. We propose that this is due to deposition of carcinoid tissue on the chordae tendineae and papillary muscles causing thickening and shortening and thence traction on the valve leaflets as the right ventricular configura-

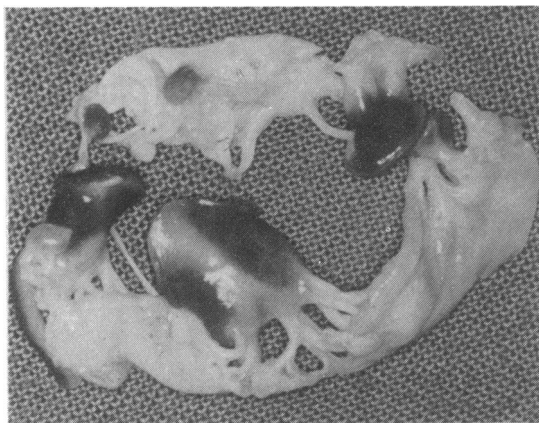


Fig. 2 Excised tricuspid valve removed at operation showing irregular and gross thickening with retraction of the leaflet.

tion changes during systole. Such a mechanism may well account for the severe tricuspid regurgitation often seen in carcinoid syndrome.

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